INVENTOR SEARCH

=> d l5 ibib abs 1

L5 ANSWER 1 OF 1 HCAPLUS COPYRIGHT 2009 ACS on STN
ACCESSION NUMBER: 2004:263765 HCAPLUS Full-text
DOCUMENT NUMBER: 140:399885
TITLE: Ascorbic acid treatment corrects the phenotype of a mouse model of Charcot-Marie-Tooth disease
AUTHOR(S): Passage, Edith; Norreel, Jean Chretien; Noack-Fraissignes, Pauline; Sanguedolce, Veronique; Pizant, Josette; Thirion, Xavier; Robaglia-Schlupp, Andree; Pellissier, Jean Francois; Fontes, Michel
CORPORATE SOURCE: Faculte de Medecine de la Timone, IPHM, Institut National de la Sante et de la Recherche Medicale UMR491, Marseille, 13385, Fr.
SOURCE: Nature Medicine (New York, NY, United States) (2004), 10(4), 396-401
CODEN: NAMEFI; ISSN: 1078-8956
PUBLISHER: Nature Publishing Group
DOCUMENT TYPE: Journal
LANGUAGE: English
AB Charcot-Marie-Tooth disease (CMT) is the most common hereditary peripheral neuropathy, affecting 1 in 2,500 people. The only treatment currently available is rehabilitation or corrective surgery. The most frequent form of the disease, CMT-1A, involves abnormal myelination of the peripheral nerves. Here we used a mouse model of CMT-1A to test the ability of ascorbic acid, a known promoter of myelination, to correct the CMT-1A phenotype. Ascorbic acid treatment resulted in substantial amelioration of the CMT-1A phenotype, and reduced the expression of PMP22 to a level below what is necessary to induce the disease phenotype. As ascorbic acid has already been approved by the FDA for other clin. indications, it offers an immediate therapeutic possibility for patients with the disease.
OS.CITING REF COUNT: 48 THERE ARE 48 CAPLUS RECORDS THAT CITE THIS RECORD (48 CITINGS)
REFERENCE COUNT: 35 THERE ARE 35 CITED REFERENCES AVAILABLE FOR THIS RECORD. ALL CITATIONS AVAILABLE IN THE RE FORMAT

RESULTS FROM SEARCHES IN REGISTRY, CAPLUS, MEDLINE, BIOSIS, EMBASE, AND DRUGU

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=> d que stat l21
L7             2 SEA FILE=REGISTRY ABB=ON  "ASCORBIC ACID"/CN
L8             1 SEA FILE=REGISTRY ABB=ON  "ASCORBYL PALMITATE"/CN
L9             1 SEA FILE=REGISTRY ABB=ON  "ASCORBYL DIPALMITATE"/CN
L11            1 SEA FILE=REGISTRY ABB=ON  N-ACETYLGLUCOSAMINE/CN
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                 OR ?ASCORBYL?(W)?PALMITATE? OR ?DIPALMITAT?(W)L(W)?ASCORBAT?
L13        20907 SEA FILE=HCAPLUS ABB=ON  L12 AND (?GLYCO? OR ?MANNO? OR
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                 2-O-ALPHA-D-GLYCOPYRANOSYL ASCORBIC ACID OR 6-O-BETA-D-GALACTOP
                 YRANOSYL L-ASCORBIC ACID OR MAGNESIUM ASCORBYL PHOSPHATE)/CN
L15          853 SEA FILE=HCAPLUS ABB=ON  L14
L16        21336 SEA FILE=HCAPLUS ABB=ON  L13 OR L15
L17            5 SEA FILE=HCAPLUS ABB=ON  L16 AND (?CHARCOT?(3A)?MARIE?)(4A)(?TO
                 OTH? OR ?TEETH? OR ?DENT?)
L18            5 SEA L17
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L20            2 SEA L19 AND (PRD<20020716 OR PD<20020716)
L21            8 SEA L19 OR L20


=> d ibib abs l21 1-8

L21  ANSWER 1 OF 8  HCAPLUS  COPYRIGHT 2009 ACS on STN
ACCESSION NUMBER:         2008:283542  HCAPLUS  Full-text
DOCUMENT NUMBER:          148:326169
TITLE:                    Generation of human embryonic stem cell-derived glial
                          and neuronal cells, and use for the treatment of CNS
                          diseases
INVENTOR(S):              Revel, Michel; Chebat, Judith; Izrael, Michal;
                          Kaufman, Rosalia
PATENT ASSIGNEE(S):       Yeda Research and Development Co. Ltd., Israel
SOURCE:                   PCT Int. Appl., 72pp.
                          CODEN: PIXXD2
DOCUMENT TYPE:            Patent
LANGUAGE:                 English
FAMILY ACC. NUM. COUNT:   1
PATENT INFORMATION:

     PATENT NO.           KIND   DATE        APPLICATION NO.         DATE
     ---------------      ----   --------    --------------------    --------
     WO 2008026198         A2    20080306    WO 2007-IL1026          20070815
     WO 2008026198         A3    20090507
         W:  AE, AG, AL, AM, AT, AU, AZ, BA, BB, BG, BH, BR, BW, BY, BZ, CA,
             CH, CN, CO, CR, CU, CZ, DE, DK, DM, DO, DZ, EC, EE, EG, ES, FI,
             GB, GD, GE, GH, GM, GT, HN, HR, HU, ID, IL, IN, IS, JP, KE, KG,
             KM, KN, KP, KR, KZ, LA, LC, LK, LR, LS, LT, LU, LY, MA, MD, ME,
             MG, MK, MN, MW, MX, MY, MZ, NA, NG, NI, NO, NZ, OM, PG, PH, PL,
             PT, RO, RS, RU, SC, SD, SE, SG, SK, SL, SM, SV, SY, TJ, TM, TN,
             TR, TT, TZ, UA, UG, US, UZ, VC, VN, ZA, ZM, ZW
         RW: AT, BE, BG, CH, CY, CZ, DE, DK, EE, ES, FI, FR, GB, GR, HU, IE,
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IS, IT, LT, LU, LV, MC, MT, NL, PL, PT, RO, SE, SI, SK, TR, BF, BJ, CF, CG, CI, CM, GA, GN, GQ, GW, ML, MR, NE, SN, TD, TG, BW, GH, GM, KE, LS, MW, MZ, NA, SD, SL, SZ, TZ, UG, ZM, ZW, AM, AZ, BY, KG, KZ, MD, RU, TJ, TM, AP, EA, EP, OA

EP 2064319 A2 20090603 EP 2007-790074 20070815

R: AT, BE, BG, CH, CY, CZ, DE, DK, EE, ES, FI, FR, GB, GR, HU, IE, IS, IT, LI, LT, LU, LV, MC, MT, NL, PL, PT, RO, SE, SI, SK, TR, AL, BA, HR, MK, RS

PRIORITY APPLN. INFO.: US 2006-840426P P 20060828
WO 2007-IL1026 W 20070815

AB A method of generating neural and glial cells (such as oligodendrocytes) is provided. The method comprising growing human embryonic stem cells under conditions which induce differentiation of the human stem cells into the neural and glial cells, said conditions comprising the presence of retinoic acid and an agent capable of down-regulating Bone Morphogenic Protein activity. The neural and glial cells of the invention are used for the treatment of medical conditions of the CNS.

L21 ANSWER 2 OF 8 HCAPLUS COPYRIGHT 2009 ACS on STN
ACCESSION NUMBER: 2007:1334626 HCAPLUS Full-text
DOCUMENT NUMBER: 148:17527
TITLE: Chimeric C3 exoenzyme-like Rho antagonists for treating injured nervous system and cancer
INVENTOR(S): McKerracher, Lisa; Munzer, Jon Scott
PATENT ASSIGNEE(S): Can.
SOURCE: U.S. Pat. Appl. Publ., 106 pp., Cont.-in-part of U.S. Ser. No. 902,878.
CODEN: USXXCO
DOCUMENT TYPE: Patent
LANGUAGE: English
FAMILY ACC. NUM. COUNT: 7
PATENT INFORMATION:

PATENT NO.	KIND	DATE	APPLICATION NO.	DATE
US 20070270340	A1	20071122	US 2006-643940	20061222 <--
CA 2342970	A1	20021012	CA 2001-2342970	20010412
CA 2362004	A1	20021012	CA 2001-2362004	20011113 <--
CA 2367636	A1	20021012	CA 2002-2367636	20020115 <--
US 20050059595	A1	20050317	US 2004-902959	20040802 <--
US 7442686	B2	20081028		
US 20060134140	A1	20060622	US 2004-902878	20040802 <--
US 20080269120	A1	20081030	US 2007-808773	20070612 <--
WO 2008077236	A1	20080703	WO 2007-CA2265	20071212

W: AE, AG, AL, AM, AT, AU, AZ, BA, BB, BG, BH, BR, BW, BY, BZ, CA, CH, CN, CO, CR, CU, CZ, DE, DK, DM, DO, DZ, EC, EE, EG, ES, FI, GB, GD, GE, GH, GM, GT, HN, HR, HU, ID, IL, IN, IS, JP, KE, KG, KM, KN, KP, KR, KZ, LA, LC, LK, LR, LS, LT, LU, LY, MA, MD, ME, MG, MK, MN, MW, MX, MY, MZ, NA, NG, NI, NO, NZ, OM, PG, PH, PL, PT, RO, RS, RU, SC, SD, SE, SG, SK, SL, SM, SV, SY, TJ, TM, TN, TR, TT, TZ, UA, UG, US, UZ, VC, VN, ZA, ZM, ZW

RW: AT, BE, BG, CH, CY, CZ, DE, DK, EE, ES, FI, FR, GB, GR, HU, IE, IS, IT, LT, LU, LV, MC, MT, NL, PL, PT, RO, SE, SI, SK, TR, BF, BJ, CF, CG, CI, CM, GA, GN, GQ, GW, ML, MR, NE, SN, TD, TG, BW, GH, GM, KE, LS, MW, MZ, NA, SD, SL, SZ, TZ, UG, ZM, ZW, AM, AZ, BY, KG, KZ, MD, RU, TJ, TM

PRIORITY APPLN. INFO.: CA 2001-2342970 A 20010412 <--
CA 2001-2362004 A 20011113 <--
CA 2002-2367636 A 20020115 <--

US 2003-506162P	P	20030929
US 2004-902878	A2	20040802
US 2004-902959	A2	20040802
US 2002-118079	A2	20020409 <--
US 2006-643940	A2	20061222
US 2007-808773	A	20070612

AB The present invention provides novel chimeric ADP-ribosyltransferase (C3 exoenzyme)-like Rho antagonists having ability to penetrate inside target cells and inactivate Rho at low doses. In some embodiments, provided are sequences for variants of C3 fused to proline-rich transport peptide. The system for example may deliver an Rho antagonist(s) in a tissue adhesive, such as a fibrin glue, to create a delivery matrix in situ. The present invention relates to the use of chimeric C3-like Rho antagonists for promoting repair and neuron survival in injured mammalian central and peripheral nervous system and for treating or preventing cancer. OS.CITING REF COUNT: 1 THERE ARE 1 CAPLUS RECORDS THAT CITE THIS RECORD

(1 CITINGS)

L21 ANSWER 3 OF 8 HCAPLUS COPYRIGHT 2009 ACS on STN
ACCESSION NUMBER: 2006:826060 HCAPLUS Full-text
DOCUMENT NUMBER: 146:198376
TITLE: Charcot-Marie-Tooth disease: correction of the CMT-1A phenotype
AUTHOR(S): Davies, Shelley L.; Moral, Ma Angels
CORPORATE SOURCE: Prous Science, Barcelona, 08080, Spain
SOURCE: Drugs of the Future (2006), 31(6), 531-533
CODEN: DRFUD4; ISSN: 0377-8282
PUBLISHER: Prous Science
DOCUMENT TYPE: Journal
LANGUAGE: English
AB Charcot-Marie-Tooth (CMT) disease constitutes a group of prevalent hereditary, chronic and debilitating peripheral neuropathies. CMT type 1A (CMT-1A), the most common form of CMT, is autosomal dominant and is characterized by peripheral demyelination. No pharmacotherapies currently exist for CMT-1A, although identification of an underlying duplication in the gene for PMP22 (peripheral myelin protein 22), a glycoprotein expressed in myelin, has ignited the search for candidates to correct the CMT-1A phenotype. To date, these include the progesterone antagonist onapristone, the antioxidant ascorbic acid and the natural neurotrophic factor neurotrophin-3.
REFERENCE COUNT: 12 THERE ARE 12 CITED REFERENCES AVAILABLE FOR THIS RECORD. ALL CITATIONS AVAILABLE IN THE RE FORMAT

L21 ANSWER 4 OF 8 HCAPLUS COPYRIGHT 2009 ACS on STN
ACCESSION NUMBER: 2004:60301 HCAPLUS Full-text
DOCUMENT NUMBER: 140:105308
TITLE: Cyclic AMP-modulating compounds and compositions for the treatment of peripheral neuropathies, preparation thereof, and uses
INVENTOR(S): Fontes, Michel; Passage, Edith; Sangeudolce, Veronique; Noreel, Jean-Chretien
PATENT ASSIGNEE(S): Universite de la Mediterranee, Fr.; Institut National de la Sante et de la Recherche Medicale; Association Francaise Contre Les Myopathies
SOURCE: PCT Int. Appl., 26 pp.
CODEN: PIXXD2
DOCUMENT TYPE: Patent
LANGUAGE: French
FAMILY ACC. NUM. COUNT: 1
PATENT INFORMATION:

PATENT NO.	KIND	DATE	APPLICATION NO.	DATE
WO 2004006911	A2	20040122	WO 2003-FR2236	20030715
WO 2004006911	A3	20040408		
W: AE, AG, AL, AM, AT, AU, AZ, BA, BB, BG, BR, BY, BZ, CA, CH, CN, CO, CR, CU, CZ, DE, DK, DM, DZ, EC, EE, ES, FI, GB, GD, GE, GH, GM, HR, HU, ID, IL, IN, IS, JP, KE, KG, KP, KR, KZ, LC, LK, LR, LS, LT, LU, LV, MA, MD, MG, MK, MN, MW, MX, MZ, NI, NO, NZ, OM, PG, PH, PL, PT, RO, RU, SC, SD, SE, SG, SK, SL, SY, TJ, TM, TN, TR, TT, TZ, UA, UG, US, UZ, VC, VN, YU, ZA, ZM, ZW				
RW: GH, GM, KE, LS, MW, MZ, SD, SL, SZ, TZ, UG, ZM, ZW, AM, AZ, BY, KG, KZ, MD, RU, TJ, TM, AT, BE, BG, CH, CY, CZ, DE, DK, EE, ES, FI, FR, GB, GR, HU, IE, IT, LU, MC, NL, PT, RO, SE, SI, SK, TR, BF, BJ, CF, CG, CI, CM, GA, GN, GQ, GW, ML, MR, NE, SN, TD, TG				
FR 2842422	A1	20040123	FR 2002-8966	20020716
FR 2842422	B1	20060630		
CA 2492368	A1	20040122	CA 2003-2492368	20030715
AU 2003271807	A1	20040202	AU 2003-271807	20030715
EP 1526850	A2	20050504	EP 2003-753643	20030715
EP 1526850	B1	20080827		
R: AT, BE, CH, DE, DK, ES, FR, GB, GR, IT, LI, LU, NL, SE, MC, PT, IE, SI, LT, LV, FI, RO, MK, CY, AL, TR, BG, CZ, EE, HU, SK				
JP 2005537264	T	20051208	JP 2004-520788	20030715
AT 406158	T	20080915	AT 2003-753643	20030715
ES 2312804	T3	20090301	ES 2003-753643	20030715
US 20050187290	A1	20050825	US 2005-521239	20050414
PRIORITY APPLN. INFO.:			FR 2002-8966 A	20020716
			WO 2003-FR2236 W	20030715

AB The invention discloses the use of a cAMP modulator in the preparation of compns. that are intended for the prevention or treatment of peripheral neuropathies. The invention further discloses tools and kits used to prepare the compns. The cAMP modulators of the invention include a variety of vitamin C compds.

OS.CITING REF COUNT: 1 THERE ARE 1 CAPLUS RECORDS THAT CITE THIS RECORD (1 CITINGS)

REFERENCE COUNT: 6 THERE ARE 6 CITED REFERENCES AVAILABLE FOR THIS RECORD. ALL CITATIONS AVAILABLE IN THE RE FORMAT

L21 ANSWER 5 OF 8 HCAPLUS COPYRIGHT 2009 ACS on STN

ACCESSION NUMBER: 1995:723143 HCAPLUS Full-text

DOCUMENT NUMBER: 123:102794

ORIGINAL REFERENCE NO.: 123:18031a,18034a

TITLE: Pharmaceutical compositions and use thereof for treatment of neurological diseases and etiologically related symptomatology.

INVENTOR(S): Shapiro, Howard K.

PATENT ASSIGNEE(S): USA

SOURCE: PCT Int. Appl., 155 pp.
CODEN: PIXXD2

DOCUMENT TYPE: Patent

LANGUAGE: English

FAMILY ACC. NUM. COUNT: 4

PATENT INFORMATION:

PATENT NO.	KIND	DATE	APPLICATION NO.	DATE
WO 9501096	A1	19950112	WO 1994-US7277	19940628 <--
W: AU, CA, JP				
RW: AT, BE, CH, DE, DK, ES, FR, GB, GR, IE, IT, LU, MC, NL, PT, SE				

US 5668117	A	19970916	US 1993-62201	19930629 <--
AU 9472144	A	19950124	AU 1994-72144	19940628 <--
AU 692454	B2	19980611		
EP 707446	A1	19960424	EP 1994-921405	19940628 <--
R: DE, FR, GB, IT				
JP 08512055	T	19961217	JP 1994-503597	19940628 <--
PRIORITY APPLN. INFO.:			US 1993-62201 A	19930629 <--
			US 1991-660561 B1	19910222 <--
			US 1993-26617 B2	19930223 <--
			WO 1994-US7277 W	19940628 <--

AB Pharmaceutical compns. for treatment of several neurol. diseases and pathophysiol.-related symptomol. in other body tissues, including peripheral neuropathies, secondary symptomol. of diabetes, Alzheimer's disease, Parkinson's disease, alc. polyneuropathy and age-onset symptomol., as well as analogous veterinary diseases, are disclosed. Spurious pathol. chemical crosslinking of normal intracellular structures is a fundamental aspect of these neurol. diseases. Covalent bond crosslinking of protein and lipid subcellular elements appear to underlie the formation of polymerized aggregates of neurofilaments and other structural proteins, and lipofuscin. Pharmacol. intervention in some neurol. diseases using water-soluble, small mol. weight primary amines or their derivs. as oral therapeutic agents, may compete with cellular protein and lipid amine groups for reaction with disease-induced carbonyl-containing aliphatic and aromatic hydrocarbons. Primary pharmacol. agents include 4-aminobenzoic acid and derivs. thereof to facilitate kidney recognition and removal. This invention also includes oral use of nonabsorbable polyamine polymers and amine-related co-agents, such as chitosan, to covalently bind and sequester potentially toxic carbonyl compds. present in the diet, oral use of known antioxidant co-agents and related nutritional factors and use of the primary agent and co-agents in combination with known medicaments for treatment of these neurol. diseases. OS.CITING REF COUNT: 19 THERE ARE 19 CAPLUS RECORDS THAT CITE THIS RECORD (23 CITINGS)
REFERENCE COUNT: 14 THERE ARE 14 CITED REFERENCES AVAILABLE FOR THIS RECORD. ALL CITATIONS AVAILABLE IN THE RE FORMAT

L21 ANSWER 6 OF 8 EMBASE COPYRIGHT (c) 2009 Elsevier B.V. All rights reserved on STN

ACCESSION NUMBER: 2008481529 EMBASE Full-text
TITLE: Charcot-Marie-Tooth disease.
AUTHOR: Lee, Yi-Chung, Dr. (correspondence); Chang, Ming-Hon
CORPORATE SOURCE: Section of Neurology, Taichung Veterans General Hospital, No. 160, Sec. 3, Chung-Kang Road, Taichung, Taiwan, Province of China. yclee@vghtc.gov.tw
AUTHOR: Lee, Yi-Chung, Dr. (correspondence); Chang, Ming-Hon; Lin, Kon-Ping
CORPORATE SOURCE: Department of Neurology, National Yang-Ming University School of Medicine, Taipei, Taiwan, Province of China. yclee@vghtc.gov.tw
AUTHOR: Lin, Kon-Ping
CORPORATE SOURCE: The Neurological Institute, Taipei Veterans General Hospital, Taipei, Taiwan, Province of China.
SOURCE: Acta Neurologica Taiwanica, (September 2008) Vol. 17, No. 3, pp. 203-213.
Refs: 84
ISSN: 1019-6099 CODEN: ANETF5
PUBLISHER: Neurological Society R.O.C (Taiwan), 7 Chung-Shan S. Road, Taipei, 100, Taiwan, Province of China.
COUNTRY: Taiwan, Province of China
DOCUMENT TYPE: Journal; Article
FILE SEGMENT: 008 Neurology and Neurosurgery
022 Human Genetics

029 Clinical and Experimental Biochemistry
037 Drug Literature Index
FILE SEGMENT: ClinicalTrials.gov
CLINICAL TRIAL NO.: NCT00484510
LANGUAGE: Chinese
SUMMARY LANGUAGE: English; Chinese
ENTRY DATE: Entered STN: 27 Oct 2008
Last Updated on STN: 27 Oct 2008

AB Charcot-Marie-Tooth disease (CMT), also called hereditary motor and sensory neuropathy (HMSN), is the most common inherited peripheral neuropathy, comprised by a group of genetically heterogeneous disorders that share clinical characteristics of progressive distal muscle weakness and atrophy, foot deformities, distal sensory loss, and depressed tendon reflexes. It can be categorized according to its electrophysiological or pathological features, transmission patterns, age of disease onset, and molecular pathology. CMT type 1 (CMT1; MIM 118200) is a group of autosomal dominant-inherited demyelinating neuropathies with a disease onset at or after childhood. Five different subtypes have been identified based on different causative genes. Among them, CMT1A (MIM # 118220) is most common and is usually associated with a duplication of a 1.5-Mb region on chromosome 17p11.2, which includes peripheral myelin protein 22 gene (PMP22; MIM *601097). Currently, there is no cure or obviously effective disease-modifying treatment for CMT. Two potential effective therapeutic agents for CMT1A were investigated recently. One is ascorbic acid and another is neurotrophin-3 (NT-3), an important component of the Schwann cell autocrine survival loop. Early diagnosis can facilitate CMT patients to modify their life styles timely for minimizing nerve injury to delay or avoid disability. Molecular diagnosis of CMT can provide the basis for appropriate genetic counseling and further CMT research.

L21 ANSWER 7 OF 8 EMBASE COPYRIGHT (c) 2009 Elsevier B.V. All rights reserved on STN
ACCESSION NUMBER: 2008461683 EMBASE Full-text
TITLE: Experimental Therapeutics in Hereditary Neuropathies: The Past, the Present, and the Future.
AUTHOR: Herrmann, David N. (correspondence)
CORPORATE SOURCE: Department of Neurology-NMD, University of Rochester Medical Center, Rochester, NY 14642, United States. David_Herrmann@urmc.rochester.edu
SOURCE: Neurotherapeutics, (October 2008) Vol. 5, No. 4, pp. 507-515.
Refs: 51
ISSN: 1933-7213
PUBLISHER: Elsevier Inc., 360 Park Avenue South, New York, NY 10010, United States.
PUBLISHER IDENT.: S 1933-7213(08)00135-9
COUNTRY: United States
DOCUMENT TYPE: Journal; Article
FILE SEGMENT: 008 Neurology and Neurosurgery
022 Human Genetics
036 Health Policy, Economics and Management
037 Drug Literature Index
038 Adverse Reactions Titles
FILE SEGMENT: ClinicalTrials.gov
CLINICAL TRIAL NO.: NCT00484510; NCT00541164
LANGUAGE: English
SUMMARY LANGUAGE: English
ENTRY DATE: Entered STN: 10 Nov 2008
Last Updated on STN: 10 Nov 2008

AB Hereditary neuropathies represent approximately 40% of undiagnosed neuropathies in a tertiary clinic setting. The Charcot- Marie-Tooth neuropathies (CMT) are the most common. Mutations in more than 40 genes have been identified to date in CMT. Approximately 50% of CMT cases are accounted for by CMT type 1A, due to a duplication within the peripheral myelin protein 22 gene (PMP22). Mutations in the gap junction beta 1 gene (GJB1), the myelin protein zero gene (MPZ), and the mitofusin 2 gene (MFN2) account for a substantial proportion of other genetically definable CMT. Some 15% of demyelinating CMT and 70% of axonal CMT await genetic clarification. Other hereditary neuropathies include the hereditary sensory and autonomic neuropathies, the familial amyloid polyneuropathies, and multisystem disorders (e.g., lipid storage diseases and inherited ataxias) that have peripheral neuropathy as a major or minor component. This review surveys the challenges of developing effective therapies for hereditary neuropathies in terms of past, present, and future experimental therapeutics in CMT. .COPYRGT. 2008 The American Society for Experimental NeuroTherapeutics, Inc.

L21 ANSWER 8 OF 8 EMBASE COPYRIGHT (c) 2009 Elsevier B.V. All rights reserved on STN

ACCESSION NUMBER: 2005111091 EMBASE Full-text
TITLE: [Genetic defects of myelination: Molecular pathogenesis of the Charcot-Marie-Tooth disease (CMT1A)].
Genetische Defekte der Myelinbildung: Molekulare Pathogenese der Charcot-Marie-Tooth Neuropathie (CMT1A).
AUTHOR: Meyer zu Horste, Gerd (correspondence)
CORPORATE SOURCE: Abteilung Neurogenetik, Max-Planck-Inst. fur Exp. Medizin, Hermann-Rein-Strasse 3, D-37075 Gottingen, Germany. g.mzh@em.mpg.de
AUTHOR: Sereda, Michael W.
CORPORATE SOURCE: Abteilung fur Neurologie, Universitatsklinikum Gottingen, Robert-Koch-Str. 40, D-37075 Gottingen, Germany. sereda@em.mpg.de
SOURCE: Neuroforum, (Feb 2005) Vol. 11, No. 1, pp. 25-30.
Refs: 5
ISSN: 0947-0875
COUNTRY: Germany
DOCUMENT TYPE: Journal; Article
FILE SEGMENT: 022 Human Genetics
029 Clinical and Experimental Biochemistry
037 Drug Literature Index
005 General Pathology and Pathological Anatomy
008 Neurology and Neurosurgery
LANGUAGE: German
SUMMARY LANGUAGE: German; English
ENTRY DATE: Entered STN: 24 Mar 2005
Last Updated on STN: 24 Mar 2005

AB Hereditary neuropathies comprise a heterogenous group of genetic disorders of the peripheral nervous system. Among the underlying defects, the malfunction of myelin-forming Schwann cells is most common and associated with dys- and demyelination of peripheral nerves. However, clinically important is the secondary degeneration of affected axons and denervated muscle fibres, both of which underlie the characteristic muscle weakness in this disease. The most frequent hereditary neuropathy, Charcot -Marie-Tooth disease type 1A (CMT1A), is caused by a 1.5Mb genomic duplication within chromosome 17. Overexpression of a structural myelin protein gene (PMP22) contained in this region leads to demyelination and axonal loss. This has been formally proven by overexpression of PMP22 in transgenic disease models. Such models are not

only important for analysing pathomechanisms of the disease. They have also proven as invaluable tools to explore novel experimental therapies for CMT1A.

SEARCH RESULTS

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=> d his ful

     (FILE 'HOME' ENTERED AT 15:56:14 ON 25 AUG 2009)

     FILE 'HCAPLUS' ENTERED AT 15:56:22 ON 25 AUG 2009
                E FONTES MICHEL/AU
L1           47 SEA ABB=ON  "FONTES MICHEL"/AU
                E PASSAGE EDITH/AU
L2           32 SEA ABB=ON  ("PASSAGE E"/AU OR "PASSAGE EDITH"/AU)
                E SANGUEDOLCE VERONIQUE/AU
L3            3 SEA ABB=ON  ("SANGUEDOLCE V"/AU OR "SANGUEDOLCE VERONIQUE"/AU)
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L4            1 SEA ABB=ON  "NOREEL JEAN CHRETIEN"/AU
L5            1 SEA ABB=ON  L1 AND L2 AND L3
L6              ANALYZE L5 1 CT :       8 TERMS

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L8            1 SEA ABB=ON  "ASCORBYL PALMITATE"/CN
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                E ASCORBYL DIPALMITATE/CN
L9            1 SEA ABB=ON  "ASCORBYL DIPALMITATE"/CN
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                E ASCORBIC ACID MANNOSYLAT/CN
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                E MANNOSYLASCORBIC ACID/CN
                E FRUCTOSYLASCORBIC ACID/CN
                E GLYCOSYLIC/CN
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                E MANNOSYL/CN
                E GLYCOSYLATE/CN
                E GLYCOSYL/CN
                E GLYCOSYLIC/CN
                E GLYCOSYLASC/CN
                E MANNOSYL/CN
L10           0 SEA ABB=ON  GALACTOSYL/CN
                E GALACTOSYL/CN
                E N-ACETYLGLUCOSAMIN/CN
L11           1 SEA ABB=ON  N-ACETYLGLUCOSAMINE/CN

     FILE 'HCAPLUS' ENTERED AT 16:06:19 ON 25 AUG 2009
L12      125170 SEA ABB=ON  L7 OR L8 OR L9 OR ?ASCORBIC?(W)ACID? OR ?ASCORBYL?(
                W)?PALMITATE? OR ?DIPALMITAT?(W)L(W)?ASCORBAT?
L13       20907 SEA ABB=ON  L12 AND (?GLYCO? OR ?MANNO? OR ?FRUCTO? OR ?FUCO?
                OR ?GALACTOSYL? OR ?ACETYLGLUCOSAMIN? OR L11 OR ?ACETYLMURAM?
                OR ?PHOSPHORYLAT? OR ?ALKALINE?(W)?EARTH?(W)?METAL? OR
                TRANSITION?(W)?METAL? OR ?SULFATE? OR ?SULPHATE? OR ?GLUCOSID?
                OR ?GLUCOPYRANOSYL? OR ?GALACTOPYRANOSYL?)

     FILE 'REGISTRY' ENTERED AT 16:10:09 ON 25 AUG 2009

     FILE 'REGISTRY' ENTERED AT 16:10:13 ON 25 AUG 2009
L14           2 SEA ABB=ON  (ASCORBYL PHOSPHATE OR ASCORBYL SULPHATE OR
                ASCORBYL SULFATE OR ASCORBYL-2-GLUCOSIDE OR 2-O-ALPHA-D-GLYCOPY
```

RANOSYL ASCORBIC ACID OR 6-O-BETA-D-GALACTOPYRANOSYL L-ASCORBIC ACID OR MAGNESIUM ASCORBYL PHOSPHATE)/CN

FILE 'HCAPLUS' ENTERED AT 16:11:30 ON 25 AUG 2009
L15 853 SEA ABB=ON L14
L16 21336 SEA ABB=ON L13 OR L15
L17 5 SEA ABB=ON L16 AND (?CHARCOT?(3A)?MARIE?)(4A)(?TOOTH? OR ?TEETH? OR ?DENT?)

FILE 'MEDLINE, BIOSIS, EMBASE, DRUGU' ENTERED AT 16:13:07 ON 25 AUG 2009
L18 5 SEA ABB=ON L17

FILE 'HCAPLUS, BIOSIS, EMBASE' ENTERED AT 16:14:43 ON 25 AUG 2009
L19 8 DUP REMOV L17 L18 (2 DUPLICATES REMOVED)
L20 2 SEA ABB=ON L19 AND (PRD<20020716 OR PD<20020716)
L21 8 SEA ABB=ON L19 OR L20

FILE HOME

FILE HCAPLUS

FILE COVERS 1907 - 25 Aug 2009 VOL 151 ISS 9
FILE LAST UPDATED: 24 Aug 2009 (20090824/ED)
REVISED CLASS FIELDS (/NCL) LAST RELOADED: Jun 2009
USPTO MANUAL OF CLASSIFICATIONS THESAURUS ISSUE DATE: Jun 2009

HCAplus now includes complete International Patent Classification (IPC) reclassification data for the second quarter of 2009.

CAS Information Use Policies apply and are available at:

http://www.cas.org/legal/infopolicy.html

This file contains CAS Registry Numbers for easy and accurate substance identification.

The ALL, BIB, MAX, and STD display formats in the CA/CAplus family of databases have been updated to include new citing references information. This enhancement may impact record import into database management software. For additional information, refer to NEWS 9.

FILE REGISTRY
Property values tagged with IC are from the ZIC/VINITI data file provided by InfoChem.

STRUCTURE FILE UPDATES: 24 AUG 2009 HIGHEST RN 1175276-34-2
DICTIONARY FILE UPDATES: 24 AUG 2009 HIGHEST RN 1175276-34-2

New CAS Information Use Policies, enter HELP USAGETERMS for details.

TSCA INFORMATION NOW CURRENT THROUGH June 26, 2009.

Please note that search-term pricing does apply when conducting SmartSELECT searches.

REGISTRY includes numerically searchable data for experimental and predicted properties as well as tags indicating availability of experimental property data in the original document. For information on property searching in REGISTRY, refer to:

http://www.cas.org/support/stngen/stndoc/properties.html

FILE MEDLINE
FILE LAST UPDATED: 22 Aug 2009 (20090822/UP). FILE COVERS 1949 TO DATE.

MEDLINE and LMEDLINE have been updated with the 2009 Medical Subject Headings (MeSH) vocabulary and tree numbers from the U.S. National Libra of Medicine (NLM). Additional information is available at

http://www.nlm.nih.gov/pubs/techbull/nd08/nd08_medline_data_changes_2009.

On February 21, 2009, MEDLINE was reloaded. See HELP RLOAD for details.

This file contains CAS Registry Numbers for easy and accurate substance identification.

See HELP RANGE before carrying out any RANGE search.

FILE BIOSIS
FILE COVERS 1926 TO DATE.
CAS REGISTRY NUMBERS AND CHEMICAL NAMES (CNs) PRESENT
FROM JANUARY 1926 TO DATE.

RECORDS LAST ADDED: 19 August 2009 (20090819/ED)

BIOSIS has been augmented with 1.8 million archival records from 1926 through 1968. These records have been re-indexed to match current BIOSIS indexing.

FILE EMBASE
FILE COVERS 1974 TO 25 Aug 2009 (20090825/ED)

EMBASE was reloaded on March 30, 2008.

EMBASE is now updated daily. SDI frequency remains weekly (default) and biweekly.

This file contains CAS Registry Numbers for easy and accurate substance identification.

Beginning January 2008, Elsevier will no longer provide EMTREE codes as part of the EMTREE thesaurus in EMBASE. Please update your current-awareness alerts (SDIs) if they contain EMTREE codes.

For further assistance, please contact your local helpdesk.

FILE DRUGU
FILE LAST UPDATED: 25 AUG 2009 <20090825/UP>

>>> DERWENT DRUG FILE (SUBSCRIBER) <<<

>>> FILE COVERS 1983 TO DATE <<<
>>> THESAURUS AVAILABLE IN /CT <<<